CASE REPORT

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Dilemmas in diagnostics and therapy of rolandic epilepsy

Dileme u dijagnostici i terapiji rolandične epilepsije

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Abstract

Introduction. It is considered that around 20%–30% of patients of all ages and in all continents have wrong epilepsy diagnoses. Diagnostic and consequential therapeutic errors appear, most often, when an adequate diagnostics is not applied. Benign focal epilepsy of childhoods with centrotemporal spikes-rolandic epilepsy, brings very often to diagnostic and therapeutic problems because of persistence of epileptic-forms changes in an electroencephalography (EEG) recording, several years after establishment of good control over seizures. **Case report.** We presented 8.5 years-old girl, with the first and the only epileptic seizure at the age of 5, during her sleep. With a clear correlation of EEG record, benign rolandic epilepsy was diagnosed, so the therapy with valproate was introduced. There were no seizures after three years of its implementation. Because of epileptic-forms changes that still persisted in EEG record during her sleep, it was suggested to further use valproate. However, after reconsidering all circumstances it was concluded that the AED should bee slowly reduced up to its exclusion. After a complete stoppage of the therapy, the patient did not have any epileptic seizure for nine months, although EEG still remained pathologically changed during her sleep. **Conclusion.** A changed EEG record in a patient with rolandic epilepsy must not be a predictor of continuation of antiepileptic drugs therapy, after 2–3 years of successful seizures remission.

Key words:
epilepsy; child; epilepsy, rolandic; diagnosis; electroencephalography; therapeutics.

Apstrakt

Uvod. Smatra se da oko 20–30% bolesnika svih uzrasta i na svim kontinentima ima pogrešnu dijagnozu epilepsije. Dijagnostičke i, posledično, terapijske greške najčešće nastaju kada se ne sprovede adekvatna dijagnostika. Benigna fokalna epilepsija u detinjstvu sa centrotemporalnim šiljcima – rolandična epilepsija, često dovodi do dijagnostičkih i terapijskih problema zbog perzistiranja epileptiformnih promena na elektroencefalogramu (EEG zapisu), nekoliko godina nakon uspostavljanja dobre kontrole napada. **Prikaz bolesnika.** Prikazana je devojčica stara 8,5 godina, koja je u petoj godini, tokom spavanja, dobila prvi i jedini epileptički napad. Uz jasnu korelaciju sa EEG nalazima zaključeno je da se radi o benignoj rolandičnoj epilepsiji i uvedena je terapija valproatom, zahvaljujući kojoj u sledeće tri godine nije bilo napada. Zbog epileptiformnih promena koje su i dalje perzisrirale na EEG snimku, tokom spavanja, predložen je nastavak uzimanja valproata. Međutim, posle ponovnog razmatranja svih okolnosti odlučeno je da se postepeno obustavi dalja primena antiepileptika. Po ukidanju terapije devojčica nije imala niti jedan epileptični napad u narednih 9 meseci, iako je njen EEG tokom sna i dalje bio abnormalan. **Zaključak.** Izmenjen EEG nalaz kod bolesnika sa rolandičnom epilepsijom ne sme biti prediktor nastavka antiepileptičku terapije, nakon 2–3 godine uspešne remisije napada.

Ključne reči:
epilepsija; deca; epilepsija, rolandična; dijagnoza; elektroencefalografija; lečenje.

Introduction

It is considered that approximately 20%–30% of patients of all ages, in all continents, have wrong epilepsy diagnoses [1, 2]. Within most of patients the diagnosis of epilepsy can be set by a precise and detailed anamnesis of epileptic seizures. It is necessary to add an additional diagnosis to the detailed seizures anamnesis; electroencephalography (EEG) while being awake and during sleeping, a detailed cytochemical analyses of the blood and urine, and, sometimes, video EEG telemetry, and also neuroradiological methods of a neurosystem image: computed tomography (CT), magnetic resonance imaging (MRI), etc. Juvenile myoclonic epilepsy – Janz syndrome, brings very often to diagnostic and therapeutic errors, and the introduction of carbamazepine, antiepileptic drug (AED) for partial seizures, brings to deterioration in epileptic seizures [3]. A benign focal epilepsy of childhood with centrotemporal spikes (BECTS) also creates often diagnostic

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and therapy dilemmas. It is the most common one of all idiopathic benign focal epilepsies in the childhood. Prevalention goes around 16% of children with epileptic seizures that start from 1 to 15 years of life. Until the end of fifties of the last century, focal epilepsies were considered as synonym for symptomatic epilepsies. However, in 1958, the first scientific papers appeared describing clinical manifestations of the focal epilepsy with children of a regular neurological analysis, and in 1965 genetic predisposition of this disease was emphasized. Genetic researches suggest polygenetic type of inheritance with the most presented aberrations onto chromosomes 15q14, then 4p15, Xp, etc, and also opinions that etiology and a way of inheritance are much more complicated and complex than it used to be thought of, originally [4]. Epileptic seizures start most often between 5–8 years of life, and it more often affected boys (60%) than girls (40%), and they appear in the first, and stop in the second decade of life, during puberty. The seizures appear most often while sleeping or during waking up. Ictal semiology makes partial seizures with typical sensor and/or sensory-motor manifestations in oropharingeal region in the sense of unilateral paresthesis of the inner cheek sides, lips, jaws, tongue; unilateral tonic, clonic or tonic-clonic contractions including cheeks, eyelid, lips, tongue, pharyngeal and laryngeal muscles; disorders, anartryes and hypersalivation [5]. BECTS seizure usually lasts one to two minutes, and during this period the consciousness is completely kept. Besides focal seizures, in younger patients, there is often secondary generalization that is often being declared as the only seizure type, if focal seizures are of weak intensity. During seizures, there is often a complete speech inability (anarthria) or the speech is not clear enough. Postictal reversible Todd's paralysis is met within younger patients where the seizures lasted longer than several minutes, to half an hour, but there is never a persistent postictal neurological deficit. Epileptic seizures appear very seldom (less than 3–4 times annually) within 65% of patients and around 10% of patients have only one seizure during the entire duration of the disease. There is about 1% of atypical BECTS forms within children, with often presented cognitive and/or neurological deficit, that can evaluate as heavier epileptic syndromes (Landau-Kleffner syndrome, epilepsy with bilateral continuous spike-waves during slow wave sleeping). Interictal EEG features normal rhythmic activities and it characterizes centrotemporal peak of spike-waves, bi or three phases (1, 5–3 Hz). Spikes or sharp waves are characteristics of BECTS; they are a bit longer, and last longer, of the amplitude between 50–200 μV, they are usually grouped in lines of different lasting, with a repeating tendency; they also appear unilaterally, and seldom bilaterally. Ictal EEG features an appearance of monorph sequence of sharp waves or rhythmical spikes above the centrotemporal regions on the opposite side in relation to clinical manifestations side. There is no significant depression before the beginning of seizures, attenuation, or basic activities. Low epileptogenity of Roland's spikes within BECTS is shown in their easy wiping by small doses application of benzodiazepines, intravenously. BECTS treatment lasts for two years, counting from the day of the last seizure. For the children with one seizure or for most children with seldom and short night seizures, antiepileptic drug (AED) treatment is not necessary. If seizures start in early childhood, the disease period is longer, and, therefore, the treatment is extended to 8 or 10 years. Abnormality persistence in EEG is not a reason for postponing a decision on treatment interruption, since normalization of EEG appears a bit later in comparison to clinical remission. The treatment should be limited to application of only one AED, mostly carbamazepine or valproate. Within atypical BECTS forms, the application of carbamazepine is not recommended, since it improves bilateral synchronization of epileptic-form of activity and deterioration of epileptic seizures [6]. BECTS prognosis is very satisfactory, since after 16 years of life, seizures stop within 98%–99% of patients. In less than 1% of adults, who had BECTS in their childhoods, generalized tonic-clonic seizures and complex focal seizures appear [5].

Case report

The patient was a 8.5-year-old girl from a regular, first term pregnancy, of regular early psychomotor development, of a negative heredity for epileptic seizures, of febrile convulsion, traumatic and inflammatory disease of the central nervous system, and also of other heredity-degenerative disease, of a regular intellectual status. The girl had her first and only seizure during sleep in her 5th year, a simple focal seizure with secondary generalization. The mother was awaken by a sound of "choking", with unilateral face muscles clonisms, that reminded on "grimaces", difficult talk, a lot of saliva, and later on, it had come to tonic-clonic contraction of the entire body extremities. After 2–3 minutes, the seizure calmed, there was no vomiting, the talk was still unclear with saliva, and after half an hour, the situation completely became normal. EEG was done, during the following day, and also EEG during her sleep that in correlation with semiology of the night seizure pointed onto benign rolandic epilepsy. Although it was only one night seizure, the therapy by valproate syrup was applied, where three years later, there was no seizure. Daily dose of valproate was 10 mg per kg of body mass, and it was regularly corrected, according to the body weight increase during the treatment period. At her last control, interictal EEG was regular, and in EEG during her sleep, there were bilateral abnormalities kept, in the sense of increased activation of unilateral rolandic sharp waves, until the appearance of bilateral, synchronic, rhythmical, higher voltage theta waves, or spike-slow wave (S-W) complexes. Due to changed EEG result during her sleep, a decision was made to further extension of AED, with a suggestion on increase of valproate dose to 20 mg/kg. The girl's parents were not satisfied with this suggestion on further extension and increase of AED dose. There was a consultation with the other epileptologist who after reconsidering all circumstances concluded that AED should be slowly reduced up to its exclusion. Valproate was reduced to 30% each week from the entire dose, and after a complete stoppage of the AED, the patient did not have any epileptic seizures for nine months. EEG still remained pathologically changed during her sleep, while the patient did not have any seizures.

Discussion

Besides a significant improvement in diagnostics and epilepsy treatment, there are numerous errors in this process and there is a great number of patients' review where epilepsy has been wrongly set or there have been errors in therapy protocol. In this case review of the patient, with rolandic epilepsy, there was an error in the decision itself on application of AED therapy, after her first epileptic seizure. Within benign epileptic syndromes in childhood, where rolandic epilepsy also belongs to, only one epileptic seizure appears very often (around 10%–30%) and in that case, there should not be applied any AED therapy. Also, it is advisable, if the seizures are seldom, last for a short time and appear only at night, that even in this case, there should not be applied AED. In the case of the presented patient, there was no time to wait to, and AED was applied after her first night epileptic seizure. Within benign focal epileptic seizures syndrome in the childhood, AED therapy is not applied regularly, after the first epileptic seizure [5]. Application of AED after the first seizure is very common in clinical practice; however, a more rational approach means application of AED only after the second seizure, unprovoked one.

After the first error made in the treatment of the presented patient, there was another error, in the sense of the decision brought on extension of the AED therapy, after three years without epileptic seizures. Since rolandic epilepsy is followed by persistent EEG abnormalities for several months, or for several years, after a stable remission of epileptic seizures, there should have not been brought a decision on extension of AED therapy. EEG result is very often a stumbling stone during setting diagnoses, and also with the choice of epilepsy therapy. Therefore, it is necessary, very carefully, in a sophisticated way, to approach to any EEG result, and surely not exceed or underestimate its significance [6]. During therapy protocol of AED treatment of benign epileptic syndrome in childhood, it should always start with gradual stoppage of antiepileptic drugs therapy, after two years of seizures remission. New researches point out the existence of patient (28%–53%) with BECTS who significantly express neuropsychological disorders in the speech sphere, cognition and behavioural, and these disorders stop in most cases during the disease remission [7]. Concerning etiology of these disorders, there are still dilemmas today and opposite opinions, where ones reconsider that these disorders are in correlation with a number and distribution of interictal EEG paroxysms [8], and the others that this correlation is not clearly separated [9]. BECTS should be treated if the seizures are frequent, last longer, and especially if they are followed by speech, cognitive and behavioural disorders. Carbamazepine is most often applied AED in BECTS therapy, and latest research point out the results of oxcarbazepine [10] and levetiracetam application [11]. A solution of current dilemmas related to diagnostics and therapy of BECTS implements a need for new, larger, controlled studies.

Conclusion

Errors in diagnostic-therapy process of epilepsy treatment are very often and can make a serious harm and influence negatively onto the quality of life. A significant number of epilepsies and epileptic syndromes in the childhood have benign nature and are characterized by a good clinic flow and a satisfactory prognosis. One of such syndromes is benign rolandic epilepsy (BECTS), where after a two-three-year period of treatment without seizures, it is necessary to exclude antiepileptic drugs therapy, regardless of persistence of epileptic-forms changes in EEG. In rolandic epilepsy, EEG result should not use as a guide or decision on its application, either during the stoppage of antiepileptic therapy. Epilepsy is a clinical diagnosis firstly, so epileptic-forms which change EEG record must not be treated, but the patient oneself.

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